Permanent Pacemaker Implantation in Infants, Children, and Adolescents

Long-term Follow-up

JAIME BENREY, M.D., PAUL C. GILLETTE, M.D., ANTOINE T. NASSALLAH, M.D., AND GRADY L. HALLMAN, M.D.

SUMMARY Twenty-four patients in the pediatric age range who underwent implantation of a cardiac pacemaker for treatment of complete atrioventricular (A-V) block were followed for an average of five years (range 1–12 years). The etiology of the A-V block was surgical in 13 cases, congenital in nine, and acquired in two. Twenty patients had symptoms of cerebrovascular insufficiency and four had congestive heart failure. To date, 18 of the 24 patients studied are alive and well. Death occurred in six patients, five of whom had complex congenital heart defects, and one of whom had Refsum’s disease. Death probably was caused by complete heart block despite pacemaker treatment in four patients, and congestive heart failure in two. In 18 of the 24 children with disabling complete A-V block, pacemaker therapy provided relief of symptoms and prolonged life.

REMARKABLE PROGRESS has been made in the management of patients with complete atrioventricular (A-V) block by artificial cardiac pacing. After the experimental work of Callaghan and Bigelow7 in 1951 and the first clinical application by Zoll8 in 1952, Weirich et al.9 applied direct myocardial stimulation in patients with surgically induced heart block. Subsequent improvements in pacemakers have been contributed by Glenn,4 Chardack,8 Zoll,8 and Kantrowitz.7

The increasing use of cardiac pacemakers in the treatment of patients with complete A-V block is the result of several factors: 1) improved and simplified techniques of both temporary and permanent pacing;2 2) advances in technology leading to more dependable electrodes and generators;3 3) increasing clinical experience and follow-up data indicating a favorable effect on prognosis; and 4) better cardiovascular performance in patients with A-V block treated with pacemakers rather than with chronotropic agents.10

Despite the extensive use of pacemakers in adults, experience with children is small. Seven series reported in the literature have listed the indications for, and short-term follow-up of pacemakers in children.11–14 A review of our 12 years experience (average follow-up, five years; range, 1–12 years) with implantation of cardiac pacemakers in 24 children is presented.

Materials and Methods

Patients

During the 11-year period, from July 1962 through September 1973, 24 patients ranging in age from eight days to 21 years (average 9.7 years), underwent implantation of a cardiac pacemaker at the Texas Children’s Hospital, St. Luke’s Episcopal Hospital, and Texas Heart Institute. There were 14 males and ten females.

Indications for pacemaker implantation included symptoms of cerebrovascular insufficiency ranging from dizziness to syncope and convulsions in 20 patients, and signs and symptoms of congestive heart failure in four patients.

Thirteen patients had complete A-V block as a complication of surgical repair of a congenital heart defect (table 1). Nine patients had congenital complete A-V block (table 2) and in four of them this was an associated congenital cardiac defect. Complete A-V block was “acquired” spontaneously in two patients. One of these had onset of A-V block at age nine years associated with Refsum’s disease15 (chronic polyneuropathy associated with ichthyosis, deafness, and retinitis pigmentosa secondary to lipid storage). Atrioventricular block had occurred in another patient at 20 years of age during her second trimester of pregnancy.

The resting ventricular rate in the 24 patients ranged from 31 to 85, average 46 beats/min. The QRS duration and morphology was normal in seven patients; 11 had left bundle branch block and six had right bundle branch block. Four patients exhibited an unstable cardiac pacemaker, shifting from a junctional rhythm to an idioventricular rhythm.

Pacemakers

The 11 pulse generators implanted from 1962 through 1967 were each of the fixed-rate type. Beginning in 1968 the demand type became available and was used in the other 13 patients. The pacemaker was implanted epicardially in all patients in this series.

Results

Patients

The 24 patients were followed from one year to 12 years (average five years). Six patients died (25%), five of whom had associated complex congenital heart defects and one of whom had Refsum’s disease (table 3). Death was sudden and unexpected in three patients. Two patients died from heart failure despite pacing and in one patient death resulted from intractable ventricular fibrillation during the replacement of a defective pulse generator.
The nine patients with congenital complete A-V block are alive and improved by the pacemaker, as are eight of 13 patients with surgical A-V block and one patient with idiopathic A-V block which developed during pregnancy.

An example of the improvement possible is that of a 9-year-old girl with isolated congenital A-V block who was considered by her parents to be asymptomatic. During a treadmill exercise test she became exhausted after only one minute. After the pacemaker was inserted the parents noted increased physical activity and the child was able to exercise for nine minutes on the treadmill.

**Pacemakers**

The average duration of pacemaker function at the completion of the follow-up study was 703 days (range 43 to 1,725 days) (table 4). In 12 patients only one unit was implanted; five of them died but in seven the initial pacemaker was functioning at the end of the study. These seven are the most recent patients in the series.

Pacemaker malfunction was observed 28 times in 12 patients. The type of malfunction included: 1) failure to capture the ventricles, 2) sudden spontaneous loss of synchronization, 3) progressive decay of pacemaker rate, and 4) non-sensing by the pacemaker of the R waves falling during the alert period.

Failure of the impulse generator occurred 23 times, 87.5% of the failures. The fault in the generator was usually depletion of the battery. Each of the deteriorating pacemakers demonstrated a change in rate as failure occurred. Electrodyne units showed an increase in interval from 10 to 150 msec at the time of failure. Three of the malfunctioning Medtronic units demonstrated repetitive firing; fortunately in these cases the impulse was below threshold for tachyarrhythmias. Other arrhythmias included premature ventricular contractions in five patients (escape bigeminy), paroxysmal ventricular tachycardia in three, and ventricular fibrillation in two.

Wire fractures, loss of contact between wire and wire in splices and between wires and metal adaptors, and dislodgement of the electrode occurred five times, 12.5% of the late failure of pacemakers in this group of patients (table 4).

**Discussion**

Reports of permanent cardiac pacing in children have been infrequent. Several reasons have been offered to explain the paucity of such cases in the literature: 1) the in-

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**Table 1. Features of 13 Patients who Experienced Postsurgical Heart Block as a Complication of Surgical Repair of a Congenital Heart Defect**

<table>
<thead>
<tr>
<th>Pt. no.</th>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Congenital heart lesion</th>
<th>Surgery</th>
<th>Symptoms</th>
<th>Ventricular rate (beats/min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>21</td>
<td>Ebstein's disease</td>
<td>TVR</td>
<td>Syncope</td>
<td>42</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>12</td>
<td>VSD, subaortic st.</td>
<td>VSD cloa. res. memb.</td>
<td>Syncope</td>
<td>50</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>5</td>
<td>VSD (type 2)</td>
<td>VSD closure</td>
<td>Syncope</td>
<td>48</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>3</td>
<td>VSD (type 2)</td>
<td>VSD closure</td>
<td>Syncope</td>
<td>85</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>13</td>
<td>VSD (type 2)</td>
<td>VSD closure</td>
<td>Syncope</td>
<td>32</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>2</td>
<td>VSD (type 2)</td>
<td>VSD closure</td>
<td>CHF</td>
<td>46</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>20</td>
<td>Tetralogy</td>
<td>Total repair</td>
<td>Syncope</td>
<td>33</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>4</td>
<td>A-V canal</td>
<td>MVR</td>
<td>CHF</td>
<td>56</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>5</td>
<td>Tetralogy</td>
<td>Total repair</td>
<td>Syncope</td>
<td>46</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>4</td>
<td>Ostium primum</td>
<td>Patch closure</td>
<td>Syncope</td>
<td>36</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>17</td>
<td>Common ventricle</td>
<td>VSD closure</td>
<td>Syncope</td>
<td>60</td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>14</td>
<td>Aortic stenosis</td>
<td>AVR</td>
<td>Syncope</td>
<td>60</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>20</td>
<td>Ostium primum</td>
<td>ASD closure</td>
<td>Syncope</td>
<td>45</td>
</tr>
</tbody>
</table>

Abbreviations: ASD = atrial septal defect; CHF = congestive heart failure; TVR = total repair; VSD = ventricular septal defect.

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**Table 2. Features of Nine Patients with Congenital Heart Block**

<table>
<thead>
<tr>
<th>Pt. no.</th>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Congenital heart lesion</th>
<th>Surgery</th>
<th>Symptoms</th>
<th>Ventricular rate (beats/min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>14</td>
<td>M</td>
<td>13</td>
<td>TGA</td>
<td>W/C shunt</td>
<td>Syncope</td>
<td>50</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>10</td>
<td>VSD, PS</td>
<td>PM impl.</td>
<td>CHF</td>
<td>52</td>
</tr>
<tr>
<td>16</td>
<td>M</td>
<td>1</td>
<td>CTGA</td>
<td>PA banding</td>
<td>Syncope</td>
<td>52</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>3</td>
<td>VSD, PS, ASD</td>
<td>Total correction</td>
<td>Syncope</td>
<td>43</td>
</tr>
<tr>
<td>18</td>
<td>F</td>
<td>10</td>
<td>None</td>
<td>PM impl.</td>
<td>Syncope</td>
<td>50</td>
</tr>
<tr>
<td>19</td>
<td>F</td>
<td>4</td>
<td>None</td>
<td>PM impl.</td>
<td>Syncope</td>
<td>52</td>
</tr>
<tr>
<td>20</td>
<td>F</td>
<td>8 days</td>
<td>None</td>
<td>PM impl.</td>
<td>CHF</td>
<td>32</td>
</tr>
<tr>
<td>21</td>
<td>F</td>
<td>9</td>
<td>None</td>
<td>PM impl.</td>
<td>Syncope</td>
<td>41</td>
</tr>
<tr>
<td>22</td>
<td>F</td>
<td>10</td>
<td>None</td>
<td>PM impl.</td>
<td>Syncope</td>
<td>30</td>
</tr>
</tbody>
</table>

Abbreviations: ASD = atrial septal defect; CHF = congestive heart failure; CTGA = corrected transposition of the great arteries; PA = pulmonary artery; PM impl. = pacemaker implantation; PS = pulmonic stenosis; PV = pulmonary valve; TGA = transposition of the great arteries; VSD = ventricular septal defect; W/C = Waterston-Cooley shunt.
cence of complete heart block in children is lower than in adults,14 2) most patients with congenital complete heart block tend to do well and do not experience symptoms or major difficulty,15,16 and 3) long-term pacing in infants and children is difficult because of the complications that arise from the small size of the patient compared to the size of the pacemaker.17 The anticipated need for multiple operations to replace batteries or wires also discourages the use of pacemakers in children. We did not find that the pacemaker upset the family life. Our patients averaged one hospitalization for one week per two years.

The prognosis in children with complete A-V block is related to the presence or absence of associated cardiovascular defects,18 stability of the cardiac pacemaker,19 and the etiology of the disease process.

Although Stokes-Adams attacks are uncommon in children with congenital complete A-V block, in cases when they have occurred many have been fatal.20-23 According to Scarpetti and Rudolph,22 Stokes-Adams attacks are the most common mechanism of death in children with complete A-V block.

In a collective series reported by Michaelsson and Engle,24 nine of 418 (2%) patients with congenital complete A-V block died suddenly. Five deaths occurred in patients who had experienced previous episodes of dizziness or syncope but did not have a pacemaker implanted. Since all of our symptomatic patients with congenital complete A-V block who required a pacemaker are alive and well, we believe pacemakers have improved their prognosis.

Complete A-V block following surgical repair of intracardiac malformations is a serious complication. The prognosis of surgical complete A-V block is almost always poor. In a series reported by Squarcia et al.25 seven patients of 911 had persistent complete heart block after leaving the operating room, five of whom died within 24 hours after surgery and the remaining two died soon after discharge. Ten of our 13 patients with surgical complete A-V block are alive. Thus the prognosis of surgical complete A-V block can be significantly improved by the use of pacemakers.

Acquired nonsurgical complete A-V block was observed in a patient with Refsum's disease in our series who exhibited syncope. Patients with Refsum's disease are prone to sudden death,18 to our knowledge no case of complete A-V block in a patient with Refsum's disease has been documented previously.

The newborn period is a vulnerable time for death, especially in children with congenital third degree A-V block. Reports of mortality occurring in the newborn period suggest that such infants with ventricular rates of less than 50 per minute are the most likely to succumb.21-23 An example is illustrated by our youngest case, an eight-day-old newborn who was admitted to Texas Children's Hospital with a ventricular rate of 32 per minute and signs of severe congestive heart failure. After successful pacemaker implantation, the child's progress has been good. The congestive heart failure cleared promptly and subsequent development of the infant has been normal.

Initial management of the infant or child with complete heart block, in whom pacing is indicated, usually involves insertion of a transvenous intracardiac electrode into the right ventricle and stimulation of the heart from an external battery-operated pulse generator. This technique protects the patient from serious arrhythmias occurring during the administration of anesthesia.

In the management of a pacemaker which has failed, a temporary transvenous pacemaker should be placed before surgery. In a pacemaker which is threatening to fail, i.e., the

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**Table 3. Features and Cause of Death in 32 Patients**

<table>
<thead>
<tr>
<th>Pt no.</th>
<th>Sex</th>
<th>Age (yes)</th>
<th>Congenital heart lesion</th>
<th>Surgery</th>
<th>Type of heart block</th>
<th>Survival (days)</th>
<th>Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>21</td>
<td>Ebstein's d.</td>
<td>TVR</td>
<td>Surgical</td>
<td>43</td>
<td>CHF</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>13</td>
<td>TGA</td>
<td>W/C shunt</td>
<td>Congenital</td>
<td>70</td>
<td>Sudden death</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>10</td>
<td>Tetralogy</td>
<td>Total repair</td>
<td>Surgical</td>
<td>1342</td>
<td>CHF</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>12</td>
<td>VSD, subaortic stenosis</td>
<td>VSD closure, res. memb.</td>
<td>Surgical</td>
<td>466</td>
<td>Sudden death</td>
</tr>
<tr>
<td>16</td>
<td>M</td>
<td>1</td>
<td>CTPA</td>
<td>PA banding</td>
<td>Congenital</td>
<td>137</td>
<td>Sudden death</td>
</tr>
<tr>
<td>23</td>
<td>M</td>
<td>13</td>
<td>Refsum's d.</td>
<td>None</td>
<td>Acquired</td>
<td>1335</td>
<td>V. Fib.</td>
</tr>
</tbody>
</table>

**Abbreviations:** CHF = congestive heart failure; CTGA = corrected transposition of the great arteries; d. = disease; PA = pulmonary artery; res. memb. = resection of subaortic memibrane; TVR = transudal valve replacement; V. Fib. = ventricular fibrillation; VSD = ventricular septal defect; W/C = Waterston-Cooley shunt.

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**Table 4. Battery Replacement in 12 Patients**

<table>
<thead>
<tr>
<th>Pt no.</th>
<th>Battery change</th>
<th>Cause</th>
<th>Duration (days)</th>
<th>Type of pacemaker</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>1</td>
<td>PM failure</td>
<td>320</td>
<td>Electrodyne</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>PM failure</td>
<td>452</td>
<td>Atricor</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>Runaway PM</td>
<td>538</td>
<td>Electrodyne</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>Broken wire</td>
<td>710</td>
<td>Atricor</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>Infection</td>
<td>243</td>
<td>Atricor</td>
</tr>
<tr>
<td>6</td>
<td>6</td>
<td>PM failure</td>
<td>464</td>
<td>Atricor</td>
</tr>
<tr>
<td>7</td>
<td>7</td>
<td>PM failure</td>
<td>148</td>
<td>Medtronic</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>PM failure</td>
<td>790</td>
<td>Medtronic</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>PM failure</td>
<td>760</td>
<td>Gen. Electric</td>
</tr>
<tr>
<td>10</td>
<td>1</td>
<td>Infection</td>
<td>373</td>
<td>Medtronic</td>
</tr>
<tr>
<td>11</td>
<td>1</td>
<td>PM failure</td>
<td>790</td>
<td>Electrodyne</td>
</tr>
<tr>
<td>12</td>
<td>2</td>
<td>PM failure</td>
<td>573</td>
<td>Electrodyne</td>
</tr>
<tr>
<td>13</td>
<td>3</td>
<td>PM failure</td>
<td>540</td>
<td>Medtronic</td>
</tr>
<tr>
<td>14</td>
<td>4</td>
<td>PM failure</td>
<td>816</td>
<td>Medtronic</td>
</tr>
<tr>
<td>24</td>
<td>1</td>
<td>PM failure</td>
<td>828</td>
<td>Cordis</td>
</tr>
<tr>
<td>25</td>
<td>2</td>
<td>PM failure</td>
<td>930</td>
<td>Medtronic</td>
</tr>
<tr>
<td>26</td>
<td>3</td>
<td>PM failure</td>
<td>1020</td>
<td>Nuclear PM</td>
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<td>12</td>
<td>1</td>
<td>Runaway PM</td>
<td>970</td>
<td>Medtronic</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>Broken wire</td>
<td>122</td>
<td>Medtronic</td>
</tr>
<tr>
<td>13</td>
<td>3</td>
<td>Runaway PM</td>
<td>1285</td>
<td>Gen. Electric</td>
</tr>
<tr>
<td>13</td>
<td>1</td>
<td>Broken wire</td>
<td>1138</td>
<td>Medtronic</td>
</tr>
<tr>
<td>23</td>
<td>1</td>
<td>PM failure</td>
<td>1535</td>
<td>Medtronic</td>
</tr>
</tbody>
</table>

PM = pacemaker.
rate is decreasing, it is usually not necessary to use a temporary pacemaker. We have found that by using regular check-ups in a pacemaker clinic, and telephone checks, emergency pacemaker changes can be largely avoided.

The results of our 12-year-experience with long-term pacing in 24 children with complete A-V block are good, not only in extending life but also in relieving symptoms. Because of the increasing reliability of the generator units, we believe a permanent cardiac pacemaker should be used in the young patient who has symptoms.

Based on the above data and those from the literature, we have developed the following criteria for implantation of a permanent pacemaker in children, of which any one of the following is an indication for implantation:

1) Surgical A-V block after two weeks
2) Stokes-Adams attack in any patient with complete A-V block
3) Congestive heart failure in a patient with complete A-V block
4) Moderately severe exercise intolerance
5) A ventricular rate less than 50 beats/min in a newborn
6) An atrial rate greater than 150 beats/min in a newborn.

References

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